## **ABSTRACT**

The invention relates to the use of tetrahydrobiopterine and the derivatives thereof in the production of a medicament to improve protein tolerance for the treatment of diseases arising from an amino acid metabolic disorder, e.g. hyperphenylalaninemia. The invention also relates to a composition which contains tetrahydrobiopterine or derivatives thereof in addition to a special mixture of amino acids. The invention can, for instance, be used as a food which is low in phenylalanine in the complete nutrition of hyperphenylalaninemic patients. Tests carried out within the text of said invention revealed that by treating the context of said invention revealed that by treating patients who had phenylalanine concentrations of more than 200 µmol/l in their blood with tetrahydrobiopterine, it was possible to reduce the concentrations of phenylalanine by 37% to 92%.